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ORIGINAL ARTICLE

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Optimum age for performing Fontan operation in patients with univentricular heart

Emmanuel Akintoye MD, MPH[1] | Gruschen R. Veldtman MD[2] | William R. Miranda MD[3] | Heidi M. Connolly MD[3] | Alexander C. Egbe MD, MPH[3]

[1]Department of Internal Medicine, Wayne State University/ Detroit Medical Center, Detroit, Michigan

[2]Department of Pediatrics, Cincinnati Children's Hospital, Ohio

[3]Department of Cardiovascular Medicine, Mayo Clinic, Rochester, Minnesota

Correspondence
Alexander Egbe, MD MPH, FACC, Mayo Clinic and Foundation, 200 First Street SW, Rochester, MN 55905.
Email: egbe.alexander@mayo.edu

Abstract

The purpose of this study was to determine the optimal age for performing Fontan operation using data from the National Inpatient Sample. Our results showed that although the Fontan operation was most commonly performed at age 2 in the United States, age 3 is the optimum age for this procedure as evident by lower rate of in-hospital mortality, procedure-related complications, and rate of nonroutine home discharge when procedure is performed at age 3 years.

1 | INTRODUCTION

The Fontan operation was initially described as a palliative procedure for patients with tricuspid atresia but the demographics of the patients undergoing this procedure has changed over time.[1] The purpose of this study was to determine the optimum age for performing Fontan completion that is associated with the lowest in-hospital mortality and postoperative complications.

2 | METHODS & RESULTS

Data were from the National Inpatient Sample (NIS) which comprise 20% stratified sampling of nationwide hospital discharges in the United States. Using in-hospital mortality as our primary end point, we evaluated the risk associated with age at procedure using a semiparametric multivariable-adjusted restricted cubic spline model. In addition, we compared risk-adjusted rates of procedure-related complications, resource utilization, and disposition at discharge between ages 0 and 10. Covariates adjusted for include gender; presence of diabetes, hypertension, congestive heart failure, chronic lung disease, renal failure, chronic liver disease, cerebrovascular disease, fluid and electrolyte disorder; hospital characteristics including teaching status and bed size; and year of procedure. We identified a total of 3319 Fontan operations performed between 2001 and 2014. The patients were 39.8% female and 52.6% whites, and the most common age at procedure was 2 (31.9%) (Figure 1). Other common age categories at Fontan procedure (in decreasing order) were 4 years (11.3%), 3 years (21.1%), 1 year (6.3%), and <1 year (9.2%); and overall rate of in-hospital mortality was 2.9%. When we evaluated the association between age at procedure and in-hospital mortality, we found a U-shaped relationship (Figure 2) with the lowest risk when procedure was performed at age 3 (risk-adjusted rate = 0.93% [95% CI = 0.23%-1.6%]). Risk-adjusted rates for other common ages (in increasing order) were 1.4% [0.24%-2.5%] at 4, 1.5% [0.77%-2.1%] at 2, 2.1% [0.19%-4.1%] at 1, and 6.1% [3.3%-9.0%] at <1 year. An increasing risk-adjusted rates of in-hospital mortality was generally observed with increasing age above 3, and ranged from 2.6% to 8.4% for procedures performed after age 10 (not shown). When we evaluated for procedure-related complications, lowest rates of sepsis occurred when procedures were performed at ages 3 (1.7% [0.73%-2.6%]) and 4 (0.90% [0.01%-1.8%]), vs national average of 3.3% [2.7%-3.9%] for a Fontan procedure; mechanical circulatory support at ages 3 (0.62% [0.1%-1.2%]), 4 (0.41% [0.01%-1.0%]), and 6 (0.39% [0.01%-1.5%]), vs national average of 1.7% [1.3%-2.3%] respiratory complications, when the procedure was performed at age 3 (12% [9.5%-14.5%]), was close to the national average (11.8% [10.8%-13%]) and at age 2 (11.4% [9.4%-13.4%]), but respiratory complication was lowest at age <1 year (3.9% [1.5%-6.3%]). We did not find significant differences between age for other complications, including hemorrhage-requiring transfusion, cardiac, vascular, neurological, venous thromboembolism, or interval pacemaker insertion. Median (Q1, Q3)

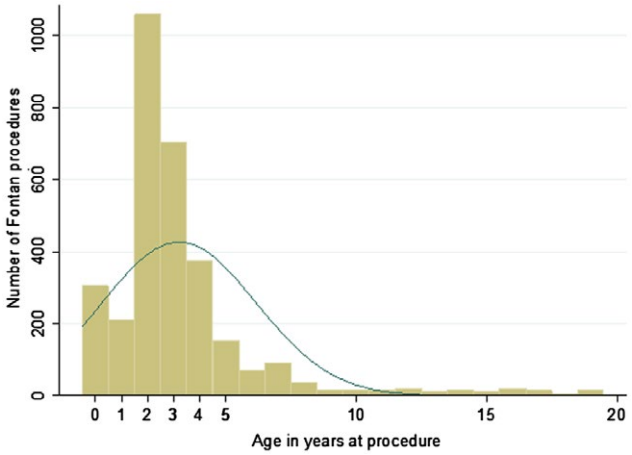

FIGURE 1 Age distribution at Fontan operation

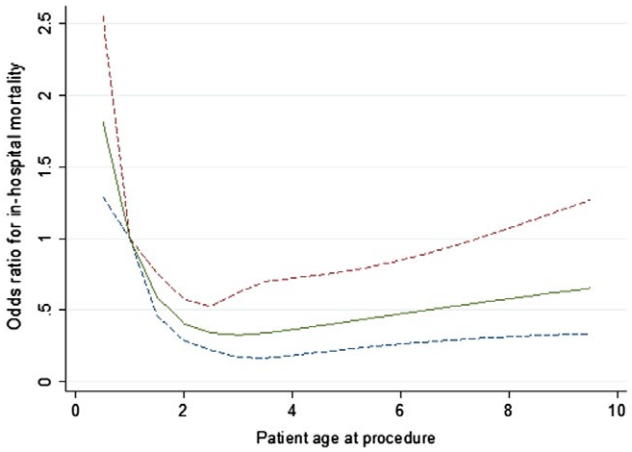

FIGURE 2 Association between age at Fontan procedure and risk of in-hospital mortality, evaluated by restricted cubic splines. The solid green line and the dashed lines represent odds ratio and 95% CI, respectively, in comparison to the reference level (age = 1). *P* value for nonlinearity <.001. The risk of in-hospital mortality continues to increase for age >10 (not shown)

hospitalization cost and length of stay at age 3 were also similar to the national average: $46 651 ($34 248.72, $72 159.55) vs national average of $46 528 ($33 962, $72 220) and 9 (7, 15) days vs 9 (7, 15) days, respectively. Lastly, the rate of nonroutine home discharge (ie, home with health care, transfer to other acute or extended care facility) was lowest for ages 3 (5.5% [3.8%-7.3%]), 4 (4.1% [2.1%-6.1%]), and 6 (4.6% [0.01%-9.3%]), vs national average of 8% [7.1%-9.0%].

3 | CONCLUSION

In summary, although the Fontan operation is most commonly performed at age 2 in the United States., our analysis of the largest nationally representative database indicates that age 3 is the optimum age for this procedure as evident by lower rate of in-hospital mortality, procedure-related complications, and rate of nonroutine home discharge when procedure is performed at age 3 years. The advantage of earlier Fontan completion is that it limits the deleterious effects of ventricular volume overload and cyanosis but the disadvantage is that it increases the odds of suboptimal Fontan hemodynamics due to inadequate development of the pulmonary vasculature resulting in high impedance to pulmonary blood flow.[2] The results of the current study is concordant with a prior single center study showing most Fontan completions were performed around the age of 2 years the current era.[3] The identification of the optimum timing for Fontan completion is novel, and will be very important in guiding congenital cardiologists and surgeons in deciding the timing of surgery that provides the best outcomes in the short run and potentially in the long term. The NIS is an administrative database and as a result some variables such as gestational age, weight, surgical techniques, and postoperative hemodynamics were not analyzed.

AUTHORS' CONTRIBUTIONS

Conception/design, data collection, analysis, manuscript drafting, critical review, and final approval: Emmanuel Akintoye; Gruschen R. Veldtman; William R. Miranda; Heidi M. Connolly; Alexander C. Egbe.

Data analysis, manuscript drafting, critical review, and final approval: Emmanuel Akintoye; Gruschen R. Veldtman; William R. Miranda; Heidi M. Connolly; Alexander C. Egbe.

ORCID

Alexander C. Egbe http://orcid.org/0000-0002-0097-6834

REFERENCES

1. Pundi KN, Johnson JN, Dearani JA, Pundi KN, et al. 40-year follow-up after the Fontan operation: long-term outcomes of 1,052 patients. *J Am Coll Cardiol*. 2015;66:1700-1710.
2. Gewillig M, Brown SC. The Fontan circulation after 45 years: update in physiology. *Heart*. 2016;102:1081-1086.
3. Murphy MO, Glatz AC, Goldberg DJ, et al. Management of early Fontan failure: a single-institution experience. *Eur J Cardiothorac Surg*. 2014;46:458-464; discussion 464.